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"SALMONELLA PARA A INFECTION IN A CASE OF PAROXYSMAL NOCTURNAL HEMOGLOBINURIA".

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INTRODUCTION:.

P.N.H. is distinctive among hemolytic disorders in man, because it is an acquired interacorpuscular defect (1). In the case described here the combination of P.N.H. and salmonella para A infection resulted in a sever-bleeding diathesis and relatiuely prolonged illness.

CASE REPORT:

A 34 old woman in march 1977 was admitted to hospital with pancytopenia (WBC-3,500 mm^3, platelets 60,000, Hb7,5 g/100) gross hemoglobinuria and hemosiderinuria. There was no history of recent drug using. Bone marrow aspiration confirmed an aplastic anemia(desert bone marrow). Diagnosis of P.N.H was estabilished by a positive acid hemolysis test. The patient treated with blood transfusion and androgens.

In April 1979 she was admitted to hospital again with a history of fever, mild jaundice, and artheralgia. On clinical examination the temperature was 39,5 and two

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days after admission she developed hepatosplenomegaly, skin rash, and diarrhea. Stool and bone marrow cultures were positive for salmonella pare A and widal seroreaction for somatic antigen was 1/80 and a week later 1/640,Hb 7 g/100cc,platelet count 25,000. Total bilirubin 4,5 mg/100,direct bilirubin 3 mg/100cc,SGot 200,SGpt 250,LDH 2,000,serum alkaline phosphatase 60 IU, absence of circulating HBsAg and a normal prothrombin time. Treatment with ampicillin was commenced. A week after admission she developed a severe generalized petechia and purpura. Increasing of ampicillin up to eight grams daily, administration of prednisolone started. Hb fell to 4 g/100cc and platelets to 15,000.

The patient was transfused with two units of whole blood. New skin hemorrhagic rash continued to appear.

She developed mental dullness and confusion. L.P was done, but no pathologic changes were seen in C.S.F. After two weeks she began to improve, fever subsided and eventually jaundice and skin hemmorrhagic rash disappeared. But her acid hemolysis test still was positive.

DISCUSSION:

P.N.H is a very rare abnormal condition. The most important test relevant to the diagnosis of hemolytic anemia is the acidified-serum test (Ham's tes)used in the diagnosis of P.N.H. The aim of this simple test is to see wether the patient's corpuscles undergo rapid hemolysis at 37c in normal serum acidified to a pH,between 6,5 and 7.0(2.3). A positive test appears to be specific for the P.N.H erythrocyte abnormality. The amount of hemolysis can be shown to be proportional to the amount of

membrane bound c3 complement(7.8)P.N.H may be secondary manifestations of a primary bone marrow injury that is manifested initially as aplastic anemia (4).

The incidence of salmonella infections has steadily increased (1) in developing countries; especially in Iran for poor socioeconomic conditions, absence of pure water supplies, contamination of milk and uncontrolling spread of organisms from persons with active disease of healthy carriers(1,6). A large group of patients who are hospitalized because of typhoid and paratyphoid fever have some type ofunderlying diseases like;aplastic anemia, hodgkin, tuberculosis,non-hodgkin lymphoma.

Patients with sickle-cell anemia and some hemolytic disorders are susceptible to salomonella bacteremia(5,10, 11) and local pyogenic infections like pyogenic liver infections(liver abcess) (2,11).

In the patient discussed here, there were a variety hematological disturbances because of salmonella bacteremia and P.N.H, making a very severe hemorrhagic terrain (9). Therefore in case of P.N.H because of presence of predisposing factors for salmonella infections, in any fever of unknown origin should remember salmonellosis.

REFERENCES:

1. Harrison (1977) principles of internal medicine. 846 eighth ed Mcgraw-Hill, New Yourk.
2. Dacie, J,V. (1970). The hemolytic anemia 25-51 2nd ed Churchill Ltd London.
3. Rosse. W.F and Dacie J.V (1966) immune lysis of normal human and paroxysmal nocturnal hemoglobinuria red

blood cells. J.clin. invest 45-736.

4. May. Jew Frank.MM (1973) paroxysmal Nocturnal hemoglobinuria lysis induced by magnesium. N.Eng. J med 255-705.
5. Woodruff A.W. (1974) Medicine in tropics 269 churchill Ltd London.
6. Hoeprich, Paul.D. (1977) Inf. diseases 652 2nd ed Harper and Row maryland.
7. Dameshek. W (1969) Foreword and a proposal for considering paroxsymal Nocturral hemoglobinuria as a candidate Myeloproliferative disorder. Blood 33: 263.
8. Ham. Th.(1973) chronic hemolytic anemia with paroxysmal nocturnalhemglobinuria N.Eng.J.Med. 217.915.
9. Mungall, Jan P.E.(1976) Rubella in a case of P.N.H Scand.J.Ing.dis 8: No,2.
10. Jawetz.E,Melnick. J.I (1976) review of medical microbiology 208 12 th ed lange Med. Publications.
11. Sherlock. S, (1975) The liver in infectious diseases of the liver and biliary system 608 fifth ed blackwell scient. publication.